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Duplicate sources of pulmonary blood supply in pulmonary atresia with ventricular septal defect

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SUMMARY In pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries, there is a tendency for each collateral artery to be the sole supply to a particular region of lung. On injection into a collateral artery, however, "wash-out" of contrast medium by non-opacified blood from a second source is sometimes seen. Alternatively, contrast medium may faintly "wash-in" to an adjacent lobar artery supplied from a second source. "Wash-in" and "wash-out" therefore show that a duplicate blood supply exists.

To assess the importance of these phenomena, we reviewed the angiograms of 50 patients with this condition. Duplicate pulmonary blood supply not resulting from surgical shunts was found in 25 patients (50%), and in 24 of 37 patients (65%) who had selective collateral injections. In one patient two collaterals anastomosed with each other. Thirty-nine instances of duplicate supply occurred in the remaining 24. The duplicate connection was to a region of lung connected to a central pulmonary artery in 29 of 39 instances (74%).

Determination of how much of the peripheral pulmonary vasculature is connected to central pulmonary arteries greatly affects decisions about palliative and corrective surgery. These results show that unless "wash-in" and "wash-out" are specifically looked for, the information obtained from angiography may be erroneous.

In patients with congenital pulmonary atresia with ventricular septal defect, the blood supply to the lungs commonly originates from major aortopulmonary collateral arteries. These frequently anastomose end-to-end with intrapulmonary arteries.[1 2] Other collateral arteries anastomose end-to-side with pulmonary arteries at the hilum, thereby becoming connected to central, usually confluent, intrapericardial pulmonary arteries.[3-5] Thus far, we have tended to assume that the resultant pulmonary blood supply is "compartmentalised" in that one major aortopulmonary collateral artery supplies one region of lung. That region of lung may consist of anything from a fraction of a segment to the entirety of both lungs, linked by confluent central pulmonary arteries.[2 5 6]

We had noticed however that when selective injections of contrast medium are made into one major aortopulmonary collateral artery, wash-out of contrast medium by non-opacified blood is sometimes seen distal to the point of injection, suggesting that more than one source of blood supplies that region of lung.

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This study was therefore carried out in order to determine how frequently such wash-out occurs, the source of the wash-out, and to what extent its existence might affect decisions on management of such patients.

Subjects and methods

Between January 1975 and July 1980, 50 patients with congenital pulmonary atresia and ventricular septal defect with major aortopulmonary collateral arteries had 79 cineangiocardiograms at The Hospital for Sick Children, Great Ormond Street, and they form the basis of this study. Care was taken to distinguish major aortopulmonary collateral arteries, present at birth, from the acquired collateral circulation.[4 5 7]

In all but one of the 50 patients, at least one aortogram had been performed. Selective injection of contrast medium into one or more major aortopulmonary collateral arteries had been made in 37 cases, and recorded by biplane cine-angiocardiography.

Central pulmonary arteries (that is intrapericardial left and/or right pulmonary arteries) were recognised most easily when they were confluent, as this results

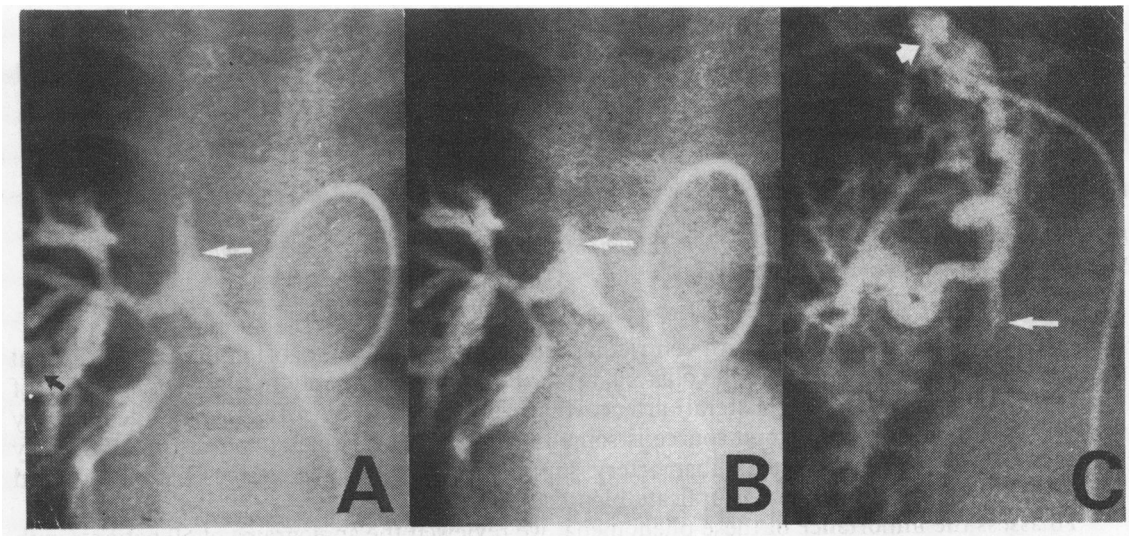

Fig. 1 *Intercollateral anastomosis (frontal projection). A and B are separate frames of the same cineangiogram of a selective injection of contrast medium into a major aortopulmonary collateral artery originating from the descending aorta. C is a separate selective injection into a collateral artery originating from the right subclavian artery and immediately bifurcating. The white arrows indicate the same point on each frame. Contrast medium running retrogradely up the vertical vessel distal to the white arrow in A is washed out in the* later *frame B. Inspection of C shows that this vertical vessel is indeed one branch of the collateral injected in C (thick white arrow), and this impression was confirmed by the wash-out occurring immediately* below *the thin white arrow, from the collateral injected in A and B.*

Note the tortuosity of vessels in A giving rise to a filling defect (black arrow), which might be interpreted as washout on a still frame. Since no swirling around this filling defect was seen when the film was run, it was not a case of wash-out.

in the "seagull" appearance in the frontal projection,[8] accentuated by head-up tilt, and the "hairpin" appearance in the lateral projection, the fusion of the two limbs of the hairpin being anterior to the trachea. Unlike major aortopulmonary collateral arteries and intrapulmonary arteries the "seagull" and "hairpin" both move predominantly with the heart rather than with the lungs. Thus, when one wing of the seagull or one limb of the hairpin is missing because of absence or atresia of the central left or right pulmonary artery, recognition of the presence of the other artery is facilitated by noting its movement with the heart. A persistent ductus arteriosus was recognised by its characteristic course, as previously described.[8]

All cineangiocardiograms, wherever the injection of contrast medium was made, were scanned by one observer (KF) for "wash-in" and "wash-out". By "wash-out" we mean that despite a constant flow of contrast medium from upstream into a vessel, non-opacified blood was seen to enter that vessel, thus washing out the contrast medium in it, and giving a swirling appearance within the vessel as contrast medium was diluted (Fig. 1–3, 4D). By "wash-in", we mean that alongside dense opacification of a region of lung opacified from injection into a given major aortopulmonary collateral artery, faint opacification of intrapulmonary arteries in an adjacent part of the lung was seen, the faintness of the opacification being the result of dilution by non-opacified blood from another source (Fig. 4D). Clearly "wash-in" and "wash-out" are but two sides of the same coin, whether one or the other is seen depends on which major aortopulmonary collateral artery is being injected.

Films thought to show "wash-in" and "wash-out" were then reviewed independently by two further observers (FJM, SGH) and "wash-in" or "wash-out" was only accepted in the case of unanimous agreement. Where "wash-in" or "wash-out" could be explained on the basis of a surgically constructed shunt, it was discounted.

When "wash-in" or "wash-out" was recognised, tracings of projected images were also made from different selective collateral injections or aortograms, or from different frames of the same angiogram, so that each collateral artery could be related to the intrapulmonary arteries with which it connected.

Results

In 49 patients (98%) the majority of major aortopulmonary collateral arteries originated directly from the aorta, usually from its descending portion,

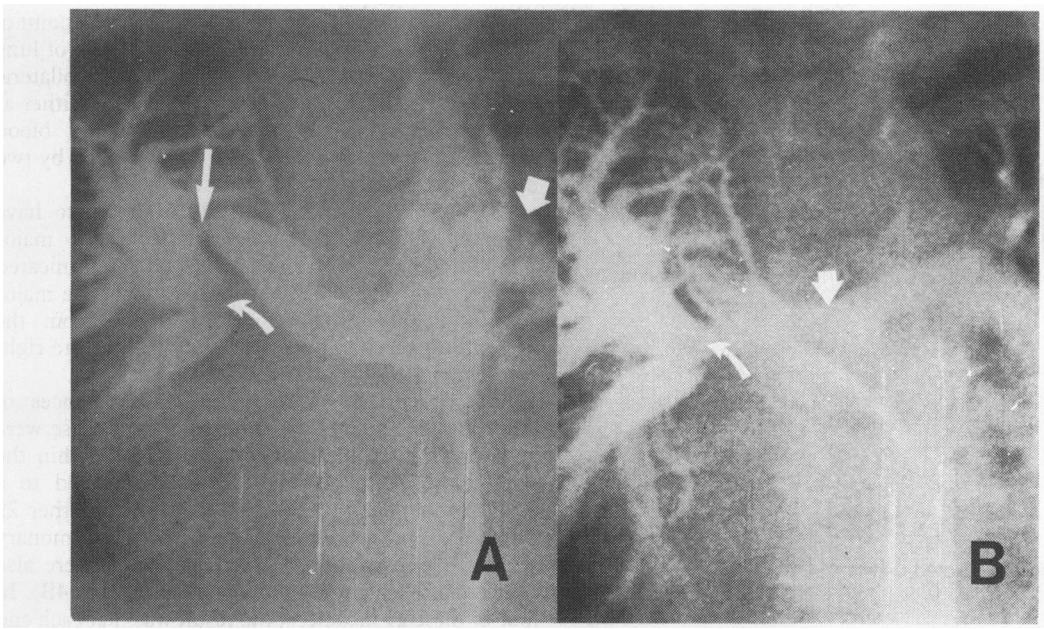

Fig. 2 *Dual supply to central pulmonary arteries (frontal projection). Two frames from the same semiselective collateral injection are shown. Early in the film (A), the right collateral is seen to anastomose with the right upper lobe pulmonary artery, the anastomosis being stenosed (straight, narrow arrow). Wash-out (curved arrow) is seen at the bifurcation of the right pulmonary artery. At this point, because of regurgitation of contrast medium into the descending aorta, faint opacification of a left collateral (straight, broad arrow) has occurred. From the left collateral (B) the confluent left and right pulmonary arteries fill (straight, broad arrow) thereby showing that the wash-out in A was originating from the central right pulmonary artery (curved white arrow).*

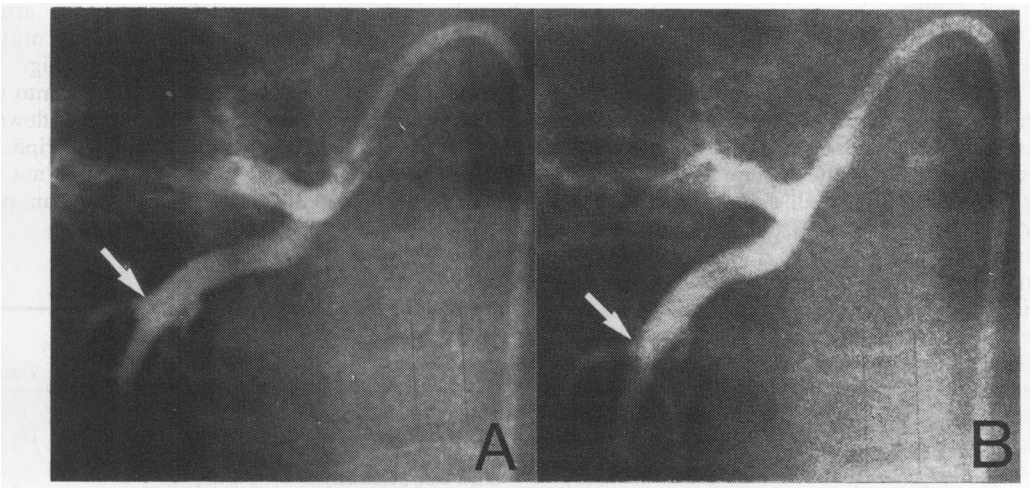

Fig. 3 *Possible retrograde flow from acquired bronchial circulation. Frontal projection of successive frames of selective collateral injection, with a white arrow marking the same point in the artery. Note that dense opacification distal to the arrow in (a) is washed out by retrograde flow in the* subsequent *frame (b).*

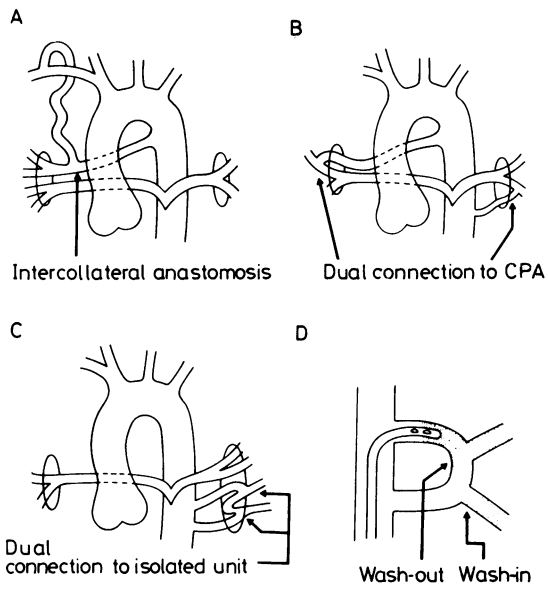

Fig. 4 *(A–C) The three main varieties of duplicate pulmonary blood supply. In C, the two arteries indicated are both supplied from two different collateral arteries. For further explanation, see text. CPA, central pulmonary artery. (D) Diagrammatic illustration of wash-in and wash-out. Two collateral arteries are linked distally by a vessel which is drawn vertically. The effect on a selective injection into the upper collateral depends on the direction of flow through the linking vessel during the injection. If that flow is upwards then wash-out is seen, as non-opacified blood enters the upper collateral and its upper continuation. If that flow is downwards then wash-in is seen as a small amount of contrast medium escapes from the upper vessels to opacify faintly the distal lower vessels.*

but in six patients one or two collaterals also arose from the underside of the aortic arch. In eight of these patients, major aortopulmonary collateral arteries also arose from the subclavian or innominate arteries, but in only one case (2%) did all major aortopulmonary collateral arteries originate from the subclavian or innominate arteries.

Central pulmonary arteries were shown in 28 of the 37 patients (76%) who had selective injections into collaterals, but in only five of 12 patients (42%) who had aortography alone. This and the subsequent surgical findings confirmed that this difference reflected the poverty of aortography as a technique for demonstrating central pulmonary arteries, rather than a real difference in incidence of absence of central pulmonary arteries in the two groups.

Wash-out was shown more frequently in patients who had selective injections of the collateral arteries, in addition to aortography, than in those who had had aortography alone (Table). No more than one point of wash-in or wash-out was seen for each region of lung connected to a given major aortopulmonary collateral artery, so we shall refer to the occurrence of either as the demonstration of duplicate pulmonary blood supply, since one region of lung was supplied by two major aortopulmonary collateral arteries.

Twenty-five patients (50%) were shown to have duplicate supply. In a single patient, two major aortopulmonary collateral arteries communicated directly with each other outside the lung, one major aortopulmonary collateral artery arising from the descending aorta and one from a branch of the right subclavian artery (Fig. 1 and 4A).

In the remaining 24 patients, 39 instances of duplicate blood supply were seen. Ten of these were between the two intrapulmonary arteries within the same lung, neither of which was connected to a central pulmonary artery (Fig. 4C). In the other 29 instances, one or both of the two intrapulmonary arteries which connected with each other were also connected to a central pulmonary artery (Fig. 4B). In four of these 29 instances, the result was that each end of the confluent central pulmonary arteries was connected to a major aortopulmonary collateral artery near the hilus (Fig. 2 and 4B). In all cases the point at which the blood supply from the two major aortopulmonary collateral arteries was seen to meet was at the hilus, or at lobar or proximal segmental artery level (Fig. 2). The frequency of duplication of blood supply was not significantly different in the right and left lungs, or between upper and lower regions of each lung.

In one patient, wash-out occurred in a segmental branch of the right lower lobe pulmonary artery which connected proximally to a major aortopulmonary collateral artery alone (Fig. 3). Furthermore, the wash-out was not coming into the side of the artery, as in all other cases, but was flowing in a retrograde manner from a more peripheral source. The origin of this wash-out could not be established, but recent experimental work in pigs

Table *Incidence of duplicate blood supply*

	No wash-in wash-out	*Wash-in or wash-out seen*	*Total*
Without aortography	1	0	1
With aortography only	11 (92%)	1 (8%)	12
Aortography + selective major aortopulmonary collateral artery injections	13 (35%)	24 (65%)	37
Total	25 (50%)	25 (50%)	50

suggests that it may have come from a true bronchopulmonary anastomosis at precapillary level (personal observations, SGH).

ASSOCIATED LESIONS

Eleven (22%) of these patients had a right aortic arch. One (2%) had transposition of the great arteries (the aorta arose from the right ventricle, and an imperforate pulmonary valve separated left ventricle from pulmonary trunk). One (2%) had a double outlet right ventricle (aorta and imperforate pulmonary valve from right ventricle). A further case (2%) had atrioventricular discordance and a further three (6%) had a persistent ductus arteriosus. In two patients, a persistent ductus arteriosus supplied one lung and major aortopulmonary collateral arteries supplied a contralateral lung, while in another patient a persistent ductus arteriosus supplied both lungs, both of which were also supplied by major aortopulmonary collateral arteries. In no case was a persistent ductus arteriosus the cause of duplicate pulmonary blood supply.

Discussion

This demonstration of duplication of pulmonary blood supply has shown that the nature of lung perfusion in pulmonary atresia is even more complex than had hitherto been imagined, and raises many unanswered questions. For example, what precisely is the relation between an arborisation abnormality, found by Alfieri and colleagues to be a major factor predicting the outcome of radical surgery,[9] and duplicate pulmonary blood supply? On the other hand, the fundamental difference between unifocal and multifocal pulmonary blood supply[5 7 10] remains unaltered. When the blood supply to the lungs is unifocal a common pressure head distributes effective flow to the entire lung. This remains true even when the pulmonary blood supply is duplicated, whether naturally, as described here, or by the creation of surgical systemic/pulmonary shunts.

The factors limiting the correctability of pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries appear to be mainly the presence and size of the central pulmonary arteries, stenoses in central and intrapulmonary arteries, pulmonary vascular obstructive disease, and the amount of lung parenchyma actually connected to the central pulmonary arteries.[2 9] An attempt at quantifying this last may be made by counting the number of pulmonary segments connected to the central vessels.[2 11] In life, this can be achieved by one of two methods. Direct evidence is selective injection into the central pulmonary artery itself, or into its source of supply.[11 12] Indirect evidence, which is inevitably less reliable, consists of selective injections into all major aortopulmonary collateral arteries not connected to central pulmonary arteries, counting the segments supplied and assuming the remainder are by default connected to the central pulmonary arteries.

Both the above methods are liable to error unless duplication of blood supply is recognised, when present. For example, let us suppose that a central pulmonary arteriogram shows seven segments (of a possible 19) connected to the central pulmonary arteries. This, on the face of it, makes the possibility of successful one-stage complete repair remote. But suppose that closer inspection reveals that there is wash-out present on this arteriogram, and that a further selective injection into the appropriate major aortopulmonary collateral artery shows dense opacification of five more segments *and* faint wash-in into an adjacent lobar artery known to be connected to the central pulmonary arteries from the pulmonary arteriogram. Then if that major aortopulmonary collateral artery is ligated and the central pulmonary artery connected to the right ventricle, 12 rather than seven segments will be perfused from the right ventricle. Provided that communication between the two systems is free (and as yet we are uncertain how to determine this), the prospect of successful surgery is brighter. This is admittedly an extreme example, but it does suggest that detection of wash-out is an important part in the preoperative assessment in these patients.

The reason why wash-out is rarely seen on aortography is obvious. Unless the injection site is such that only some collaterals are opacified, there will be no opportunity for wash-out to occur. This means that the true incidence of duplicate pulmonary blood supply is around 65%, as obtained in patients who had selective collateral injections. Even this may be an underestimate since, if in a given patient neither of the two sources of duplicate supply was injected, then duplicate supply would not be recognised. There was no patient, however, who had selective injections in whom, as far as we know, two collaterals were not injected. We suspect that wash-out on selective injection into major aortopulmonary collateral arteries could not be reliably detected except with cineangiography. This is because the tortuosity and overlapping of vessels seen in the region of the hilus frequently gives the appearance of a filling defect within a vessel on a still frame (Fig. 1). A filling defect which is suspicious of wash-out can only be identified as such when the cine film is run, and swirling of partly opacified blood round the filling defect is seen.

Thus, duplication of pulmonary blood supply probably explains why in a study of patients investigated before and after creation of a surgical shunt into a central pulmonary artery, more segments

were found connected to the central pulmonary arteries after than before operation.[11] Presumably the "missing" segments of lung were also perfused by major aortopulmonary collateral arteries but creation of the shunt encouraged blood to flow from the central pulmonary arteries into the "missing" segments rather than in the reverse direction. The arguments for unifocalisation of "compartmentalised" pulmonary blood supply remain as strong as ever, but before such an operation is carried out, a careful search for wash-out should be made before two apparently isolated lobes or segments are connected, lest they already communicate with each other.

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